Congenital Abnormalities

The Timing and Management of Surgical Reconstruction In Infancy and Childhood

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• **The proper timing of corrective and reconstructive surgical operations for the management of congenital abnormalities in infancy and childhood is exceptionally important. Although no hard and fast rules apply, each deformity should be evaluated in the light of specific features of local and general relationship. Correlation to growth and development must be stressed as an important feature. Evaluation of the congenital abnormality in the light of its many relationships provides for a basis of better complete handling, follow-up, and ultimate result with the aim directed toward elimination of tumor threat, restoration of function and esthetic improvement.**

ALTHOUGH most physicians know generally that many of the congenital abnormalities of childhood can be corrected by plastic and reconstructive surgical measures, problems having to do with the correlation of management with the proper timing of surgical correction and reconstruction are not always fully appreciated. Several important aspects of the commoner problems that confront plastic surgeons will be reviewed here to provide insight into the general plan of surgical appraisal; and some of the features of these problems will be presented by example.

Physicians trying to evaluate a defect or anomaly are broadly concerned with questions of:

- Diagnosis
- Direct effect on function and degree of anatomic distortion
- Relation to growth and development
- Possibility of secondary change or conversion to malignancy
- Methods or techniques available for management and reconstruction
- Whether to undertake correction early or wait.

The commoner problems concern lesions of two broad categories: Developmental defects and tumors of soft tissues. Under developmental defects, the incidence of cleft lip and palate deformity is 1 in 750 births. This anomaly will serve for purposes of illustrating the analysis behind the timing of surgical repair. The degree of defect can vary; it can be incomplete or complete and it can be either unilateral or bilateral. In cases of complete structural anomaly there is functional embarrassment owing to lack of continuity of orbicularis oris muscle, alveolar ridge and hard and soft palates. Functional loss is manifest by velo-pharyngeal incompetence, defective deglutition, absence of soft tissue, dynamic molding of the anterior one third of facial architecture, and—of great importance—potential loss of effective speech. To correct these abnormal features, the plastic surgeon plans to carry out structural realignment of tissue at a time when the amount of tissue is adequate and the risk of impairing further growth and development is minimal. The cleft lip deformity is best corrected shortly after birth, when the infant has regained its birth weight. In incomplete lip cleft, a delay up to three months need not have adverse effect, since the disruption of soft tissue continuity is only partial. With the use of the advanced technique of Le Mesurier and others in which measured flaps are employed and by placing atraumatic sutures and handling tissue with great care, early repair of the lip deformity can be done with good functional and cosmetic results (See Figure 1). Following the repair of the lip, growth and molding of the underlying alveolar ridge and palate will take place with narrowing of the palatal defect, which then can be surgically corrected in one or two stages between age of 18 and 24 months. This period is chosen since there will be sufficient tissue for the surgical repair and minimal chance of affecting bony growth of the mid-face. Careful detailed follow-up of the reconstructed lip and palate provide for evaluation of dentition, speech, pattern of growth, development and appearance, since at no time in early anabolic life are we facing a static situation. Minor secondary revision of the lip or nose can be considered after four years of age; and the use of a pharyngeal flap as a measure directed toward speech improvement can also be considered after evaluation of speech and velo-pharyngeal competency.

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Less common are oblique facial clefts. In the case of the patient pictured in Figure 2 there were several anomalies:

1. Oblique cleft of the face with secondary macrostomia
2. Incomplete cleft of the lip on left
3. Microtia
4. Pre-auricular appendage on left.

The lip was discontinuous at two points, so initially the cleft of the lip was repaired shortly after birth; then, after sufficient growth, the oblique facial cleft was repaired, the patient then being five and a half months old.

The reconstructive correction of ear deformities is delayed until the patient is 4 or 5 years old (the preschool period) so that development can provide for sufficient growth of rib cartilage (which is needed as an autogenous graft) and sufficient facial development to provide the soft tissue needed for ear reconstruction. This plan provides additionally for restoration of near normal symmetry to the face so that the element of dissimilarity is minimized in the school years. The small pre-auricular appendage can be removed surgically during one stage of ear reconstruction.

Among hand deformities, syndactylism can be used as an example of the use of proper timing or repair to prevent potential distortion of the digits during growth. The deformity can exist in varying degrees. Essentially, the fully developed digits are of unequal length and the epiphyseal growth (not complete in the phalanges until approximately age 16) is such that any uncorrected deformity can lead to malfunction and disfigurement. Within the first or second year of life, complete syndactylism should be corrected by web division and reconstruction, using local flaps and free full thickness skin grafts. Partial non-restricting syndactylism should also be corrected early because of the functional deficit and the potential social stigma. Figure 3 shows first the preoperative configuration of partial syndactylism involving the long and ring fingers, then the normal anatomic configuration (with normal function) that was achieved by web space reconstruction using triangular flaps and free full-thickness skin grafts.

All abnormalities of the hand must be evaluated with regard to degree of deformity, functional deficit, potential adverse effect on future growth, potential adverse social effect on family and society.

Figure 4 shows a newborn infant with supernumerary thumbs—each side presenting a different deformity. The extra right thumb, a useless segment of tissue attached by a small stalk, was surgically removed in the newborn nursery. The other thumb was a duplication with bony deformity of the phalanges. For deformity of this kind detailed bony and soft tissue reconstruction is necessary. Hence opera-

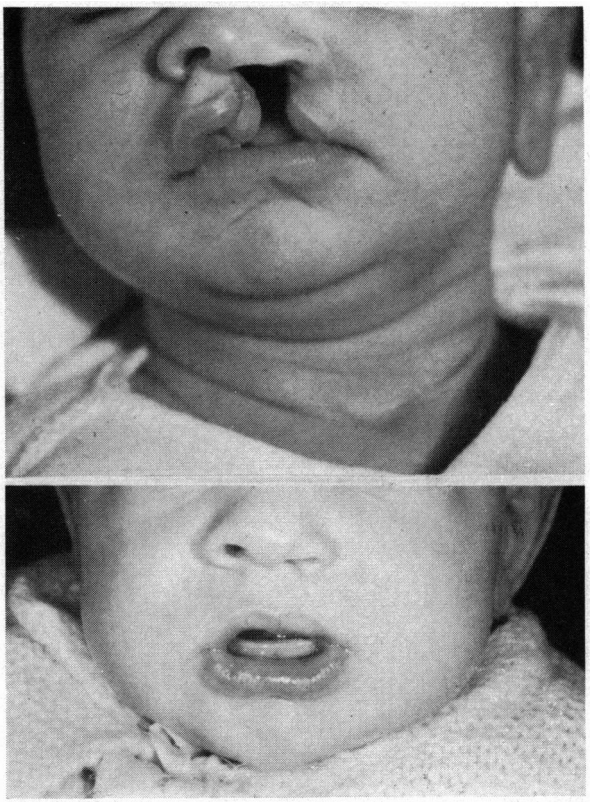

Figure 1.—Repair of lip deformity. Upper picture shows deformity at time of operation (infant 8 days old) and lower the result, the patient then 18 months of age.

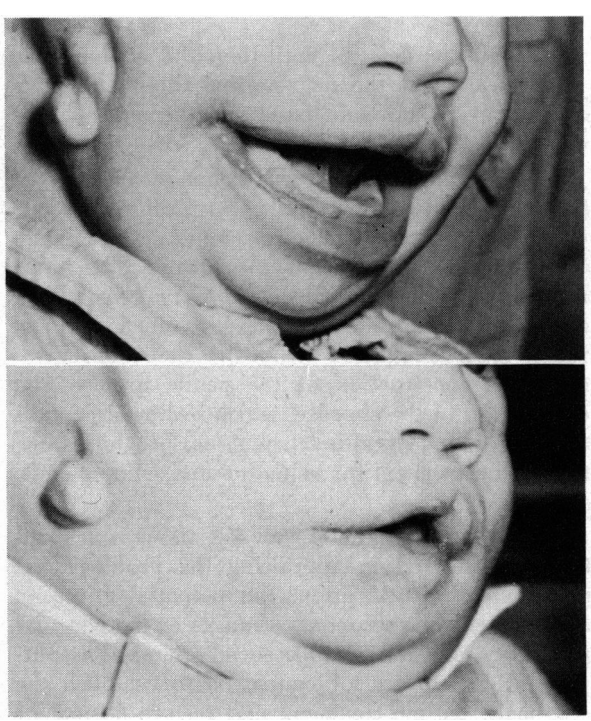

Figure 2.—Oblique facial cleft repaired when infant was five and a half months old. Reconstruction of ear to be delayed to age 4 or 5 to permit development of needed rib cartilage and soft tissue for procedure.

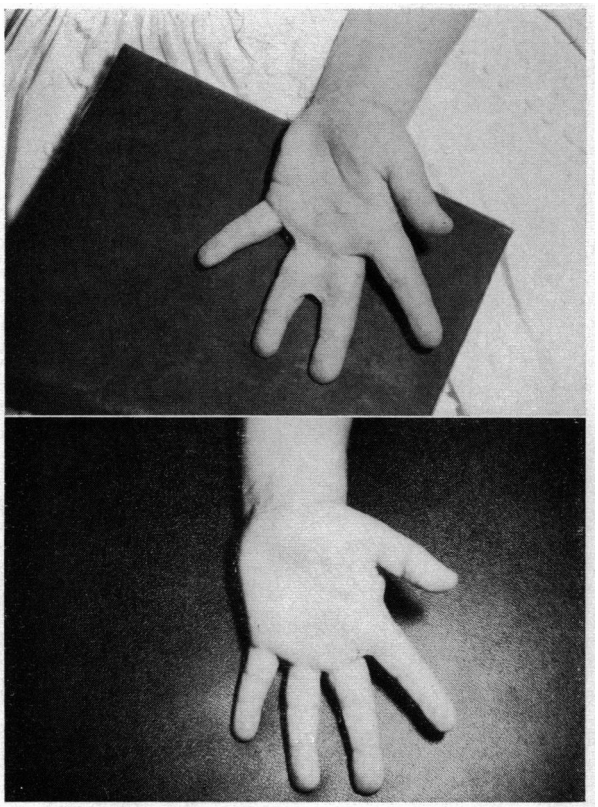

Figure 3.—The deformity above was corrected by web space reconstruction, using triangular flaps and free full-thickness skin grafts.

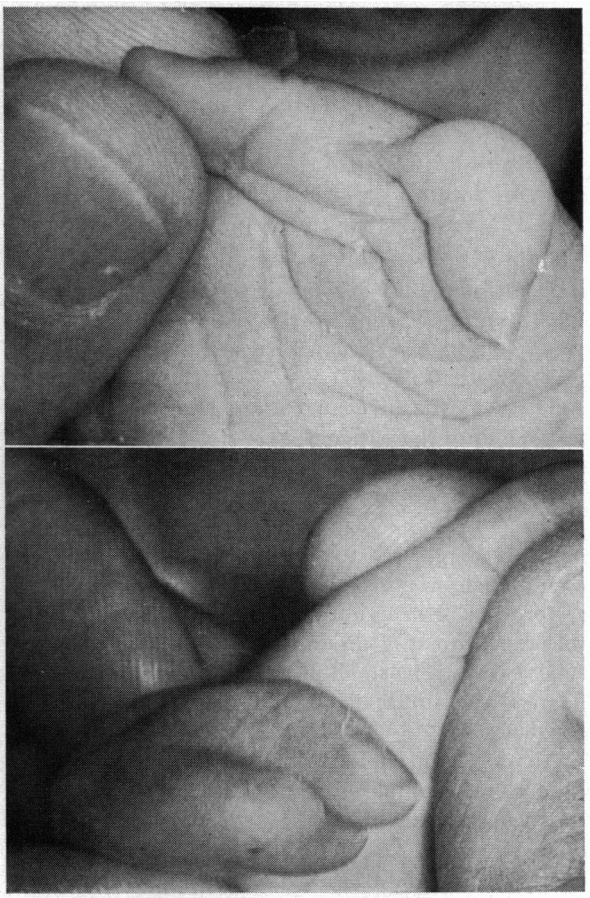

Figure 4.—*Above*, extra right thumb, attached by a small stalk, was removed in the newborn nursery. *Below*, supernumerary thumb on the left hand of the same patient. Such deformity requires detailed bony and soft tissue reconstruction and is best delayed until about a year after birth lest the operation jeopardize the near-normal thumb that is to be salvaged.

tion should be delayed until the child is about one year of age lest the near-normal thumb be jeopardized. Bony and soft tissue derangement must be evaluated on the conditions in each case.

Hypospadias is not a very common anomaly of the male genito-urinary tract. Surgical reconstruction of this deformity usually commences after sufficient growth has taken place to provide the tissue needed, yet before school period so that the stigma attached to sitting for urination is avoided. Surgical repair is accomplished ordinarily in two or three stages. Reconstruction of the penile urethra and elimination of the chordee, which ordinarily entails the use of some grafting technique, provides functional improvement for urination and seminal emission.

In dealing with tumors of soft tissue, questions to be considered in appraising the problems are microscopic identification, the potential effect on development, the secondary changes or the effect by proximity, and the possible social stigma. The purpose of all studies, of course, is to establish the diagnosis, outline treatment and restore normal or near-normal anatomic relationship.

Gynecomastia is a well recognized entity of this sort. Corrective efforts are aimed at eliminating ab-

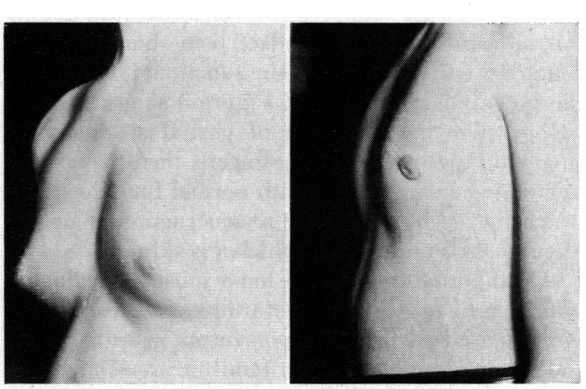

Figure 5.—Gynecomastia corrected soon after it developed, at puberty, to obviate psychic trauma.

normal breast tissue, restoring male configuration to the thorax and obviating potential psychic trauma. As soon as the deformity presents itself, at puberty, it can be corrected by properly planned excision in one stage. Figure 5 shows typical gynecomastia, un-

associated with any other deformities, in a 14 year-old boy. In this case the abnormal breast tissue was removed through subareolar incisions.

In the management of hemangiomas—whether capillary, cavernous or of mixed type—careful appraisal is necessary. Most of these "strawberry" birth marks involute spontaneously, but natural involution can be hastened or the activity controlled by the judicious injection of sclerosing agents or the application of carbon dioxide snow. Roentgen therapy entails risk of destruction of growth centers, but expertly done it may be useful in some cases. Rarely is surgical intervention needed except in the case of a rapidly advancing tumor that has not responded to simpler methods. Figure 6 shows a mixed hemangioma in the entire substance of the lower eyelid. One month after a single treatment with carbon dioxide snow, there was evidence of involution and regression as manifest by graying of the surface and definite firmness of the deeper tissue. This method is consistent with preservation of the substance of the lower eyelid.

Figure 7 shows a mixed or capillary-cavernous hemangioma in the region of the parotid gland and the seventh cranial nerve. The mass was cherry red on the surface and irregularly spongy in the deeper areas. After the use of carbon dioxide snow, the capillary component changed from bright red to gray and there was firmness of the deeper tumor, indicating involution.

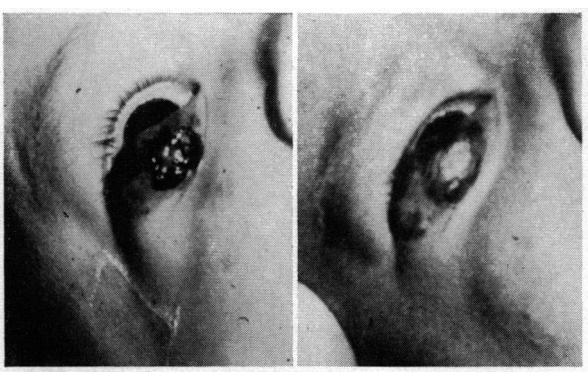

Figure 6.—*Left,* mixed hemangioma occupying entire lower lid. Right, evidence of involution a month after one treatment with carbon dioxide snow.

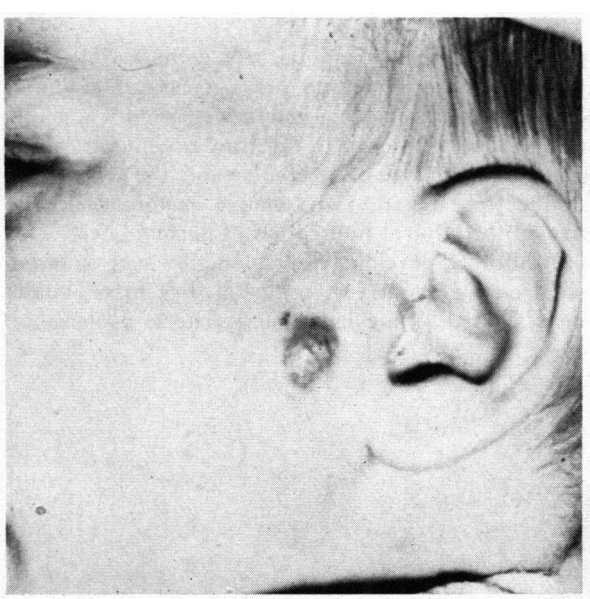

Figure 7.—Capillary-cavernous hemangioma. It was cherry red on the surface, irregularly spongy in deeper areas. After use of carbon dioxide snow the capillary component changed to gray and the deeper tumor firmed, indicating involution.

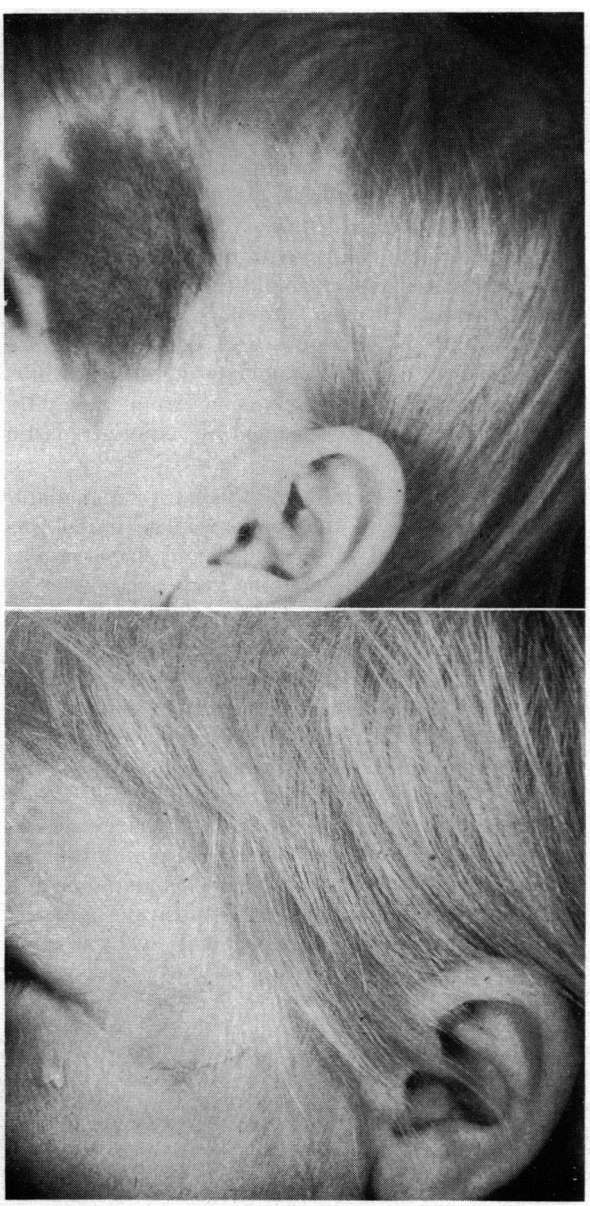

Figure 8.—A giant hairy nevus, present from birth, was surgically removed in one stage and the area covered with a free dermatome graft.

Aggressive treatment in any critical area could lead potentially to great functional and structural loss, and it is well known that conservative management in the great majority of cases offers the best results. Late in the care of hemangiomas, when all evidence of activity has disappeared, the resultant scar of remaining fibrous elements produced by involution of the deep cavernous components can be selectively dealt with by surgical excision. This reduces the cosmetic significance of lesions in the exposed areas.

Lymphangioma, on the other hand, rarely if ever undergoes spontaneous involution and, although benign, it may increase in size and produce disfigurement and changes in the adjacent tissue by expansion. The problem can be dealt with by selective surgical excision and obliteration of lymph spaces by aspiration and injection of sclerosing agents.

Pigmented nevi are commonly seen in various sizes, locations and numbers. Selected surgical correction of the larger exposed lesions is advocated both to eliminate the possibility of malignant conversion and to help cosmetically. For exposed lesions, excision in the pre-school period serves cosmetic and psychic purposes. Others should be excised before puberty to obviate the effect of hormonal change. Psychic trauma from unsightly scarring often can be prevented by judicious plastic repair.

Figure 8 shows a child who was born with a giant-type hairy nevus of the forehead. The lesion was surgically removed in one stage and the area was covered with a free dermatome graft.

Dermoid cysts, commonly seen in the eyebrow or about the nose, often warrant operation at an early age. Such cysts come about from faulty tissue fusion or isolation of a section of ectodermal elements deep within mesodermal tissue. Progressive enlargement, bone erosion and possible extension inward beyond the skull are indications for removal as well as for positive pathological identification. Presurgical evaluation should, of course, include the possibilities of glioma, neurofibroma, cephalocele, hemangioma and post-traumatic fibroma. Figure 9 illustrates a cystic, deep swelling in the right medial eyebrow area

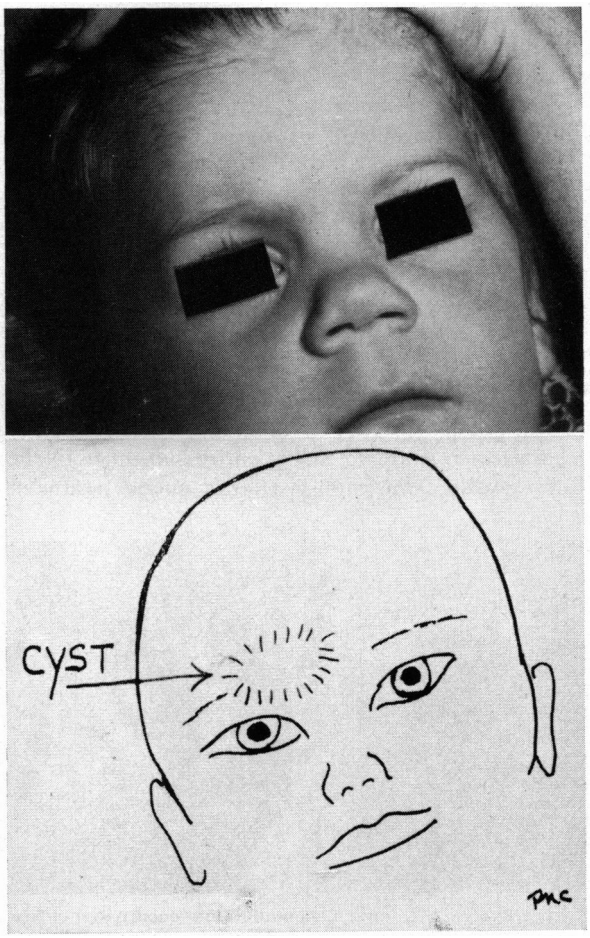

Figure 9.—Cystic swelling in left eyebrow, surgically removed.

which was slightly mobile, non-compressible, discrete and did not enlarge during crying. The tumor was surgically removed, and as suspected it was observed to extend deep to the frontalis muscle, adjacent to, but without extension beyond, the periosteum of the frontal bone. Microscopic examination confirmed the clinical impression of dermoid cyst.

Additional developmental anomalies such as branchial cleft cysts and thyroglossal duct cysts should be evaluated by much the same criteria as dermoid cysts.

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